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Case report

# Fibrous dysplasia of the temporal bone

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**Abstract**

Fibrous dysplasia is characterized by a progressive replacement of normal bone elements by fibrous tissue. It is seen in the two first decades of life and is equally distributed in relation to the sex of patients. The temporal bone is involved in 18% of the cases. Fibrous dysplasia of the temporal bone is more commonly manifested in the external auditory canal (80%) and the inner and medium ear can also be involved. Sarcomatous alterations are rare. Diagnosis is based on radiological images. Treatment is conservative and surgery is reserved for preserving function and preventing complications. The case of a male patient, 16 years old, who presented with progressive hearing loss over a 1-year period is reported. 

*Keywords:* Fibrous dysplasia; Temporal bone; Children

## 1. Introduction

The term fibrous dysplasia was described by Von Recklinghausen, although it was introduced by Lichtenstein in 1938 [1]. Fibrous dysplasia is a disease characterized by the progressive replacement of normal bone elements by fibrous tissue. Fibrous dysplasia can be classified as:

- monostotic: involving only one bone (70%);
- polyostotic: involving multiple bones (30%);
- Mc Cune Albright's syndrome: bone involvement associated with skin lesions and endocrinopathies (3%).

Craniofacial structures are involved in 10% of monostotic fibrous dysplasia cases, 50% of mild polyostotic and in 100% of severe polyostotic cases [2–4]. In this region, the most affected areas are the upper maxilla and the mandible. The temporal bone is affected in 18% of cases. There are no reported cases of Mc Cune Albright's syndrome involving the temporal bone.

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## 2. Pathophysiology

There are different theories concerning the etiology of fibrous dysplasia [5]. Fibrous dysplasia may be:

- a congenital anomaly of the mesenchyma bone forming portion, leading to pathological bone;
- a disorder of the normal repair process after an injury;
- a sequestration of maturating bone.

## 3. Histology

The stroma is formed by a collagen matrix with fibroblasts arranged in a spiral pattern [3]. Bone trabeculae have a puzzle-like co18
nfiguration. There is no invasion of the periosteum. Transition between normal and dysplastic areas of the bone cannot be distinguished as there is no true capsule in the borders of the lesion. The bone marrow and trabeculae from a soft region may be white or red, depending on the vascularization of the lesion.

## 4. Clinical picture

Symptoms result from an abnormal bone growth that can displace adjacent structures, producing compression and altered functions or cosmetic deformity.

### *4.1. Involvement of the external auditory canal*

The external auditory canal is involved in 80% of cases [6]. Progressive stenosis can lead to conductive hearing loss as well as cholesteatoma secondary to entrapped keratin. The external growth of fibrous dysplasia can lead to asymmetry and cosmetic deformity.

### *4.2. Involvement of the middle ear*

The middle ear is rarely involved and this only happens after a long period of external auditory canal stenosis. Complications of fibrous dysplasia in the middle ear include chronic otitis media with or without cholesteatoma, destruction of the ossicles, labyrinthitis, perceptive hypoacousia and facial paralysis.

### *4.3. Involvement of the inner ear*

According to some authors [7,8], fibrous dysplasia does not involve the inner ear structures; however, some cases have been reported [6] where the optical capsule was involved, affecting the cochlea and the labyrinth. Perceptive hypoacousia may occur due to cochlear destruction, stenosis of the inner auditory canal or vestibular fistula.

### *4.4. Involvement of the temporomandibular joint*

Excessive growth bones of this joint can lead to a decrease in the excursion of the mandible and to poor dental occlusion. With progression of the disease into the temporal bone, the central nervous system and/for cranial pairs may be affected. Sarcomatous alterations are rare (0.4%) [5] and can be secondary to radiation therapy [9].

## 5. Imaging studies

There are three radiological groups: pagetoid, sclerotic and cystic [3,5]:

1. pagetoid (56%): 'ground glass' is the most common form and is made up by a mixture of dense and radiolucent areas of fibrosis;
2. sclerotic (23%): these are the homogeneous dense areas;
3. cystic (21%): a radiolucent ring, oval or round, surrounded by a capsule of dense bone.

Related histology shows that the pagetoid lesions are the result of junctions between fibrous tissue and bone trabeculae. They produce radiodense and radiolucent areas, resulting in the classic ground glass appearance. The predominance of bone elements results in the opaque sclerotic type and the predominance of fibrous elements in the cystic type.

Computerized tomography is the best radiological study to show changes in the temporal bone. Nuclear magnetic resonance is used in cystic fibrous dysplasia cases. It is useful in the evalua-

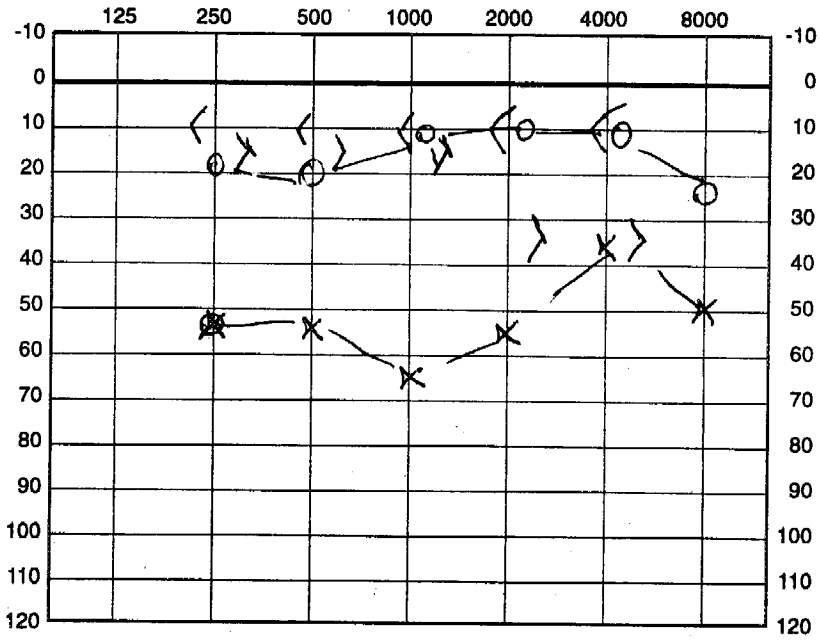


Fig. 1. Audiogram shows the conductive left hearing loss.

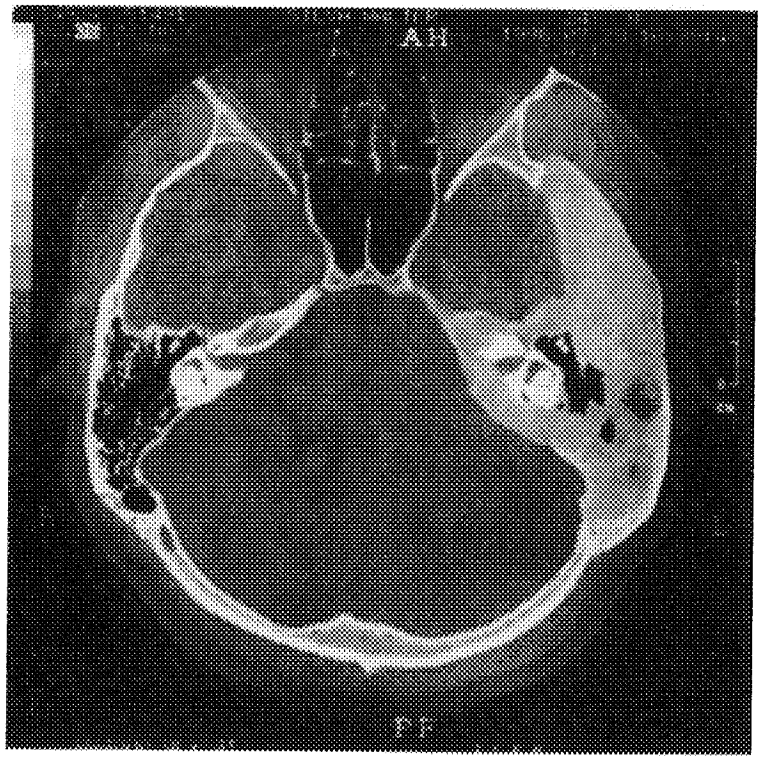

Fig. 2. CT scan: shows the left temporal bone disorder, but not affecting the middle ear cavity and labyrinth.

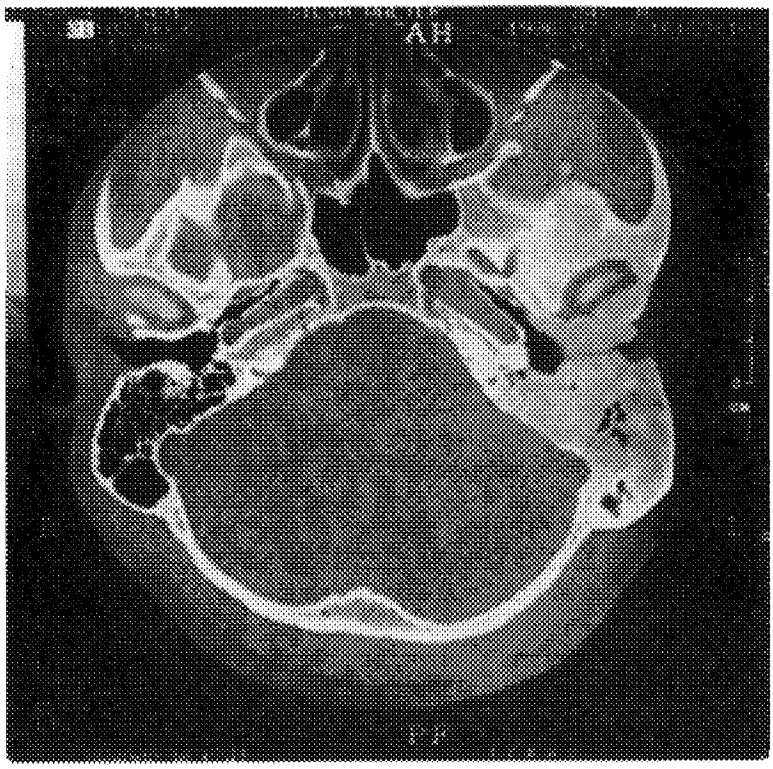

Fig. 3. Axial CT scan: shows the stenosis of the external auditory canal of the left side.

tion of soft tissues and fibrous components, and to assess the effect of these primary lesions adjacent to the soft tissue structures of the skull base, jugular vein and brain stem.

Scintillography with Tc99 m HMDP and Ga67 [10] is an imaging technique that detects and identifies polyostotic locations of fibrous dysplasia [11]. Due to the increased vascularization of these lesions, there is an active concentration of radioactive drugs in bone, both in the early and late phases.

## 6. Differential diagnoses

Differential diagnoses of osteofibrosis lesions of the temporal bone include [12]:

- Bone aneurysmatic cyst: formed by spaces filled with blood, separated from connective tissue by bone tissue or osteoid trabeculae and giant cells.
- Paget's disease: it is characterized by anarchic bone absorption, leading to a complete disorganization of the trabecular structure of the affected bones.
- Osteochondroma: bone growth lined by cartilage that is formed on the surface of the affected bone. It is due to an excessive activity of the periosteum, that tends to form abnormal foci of metaplastic cartilage.
- Osteoma: a bone neoformation by osteoblastic connective tissue forming abundant osteoid, a well differentiated tissue with laminar structure and very slow growth.

The diagnosis of dysplasia is confirmed by a combination of radiographical imaging and histological data. The presence of laminar bone tissue and a fibrous matrix suggest differential characteristics.

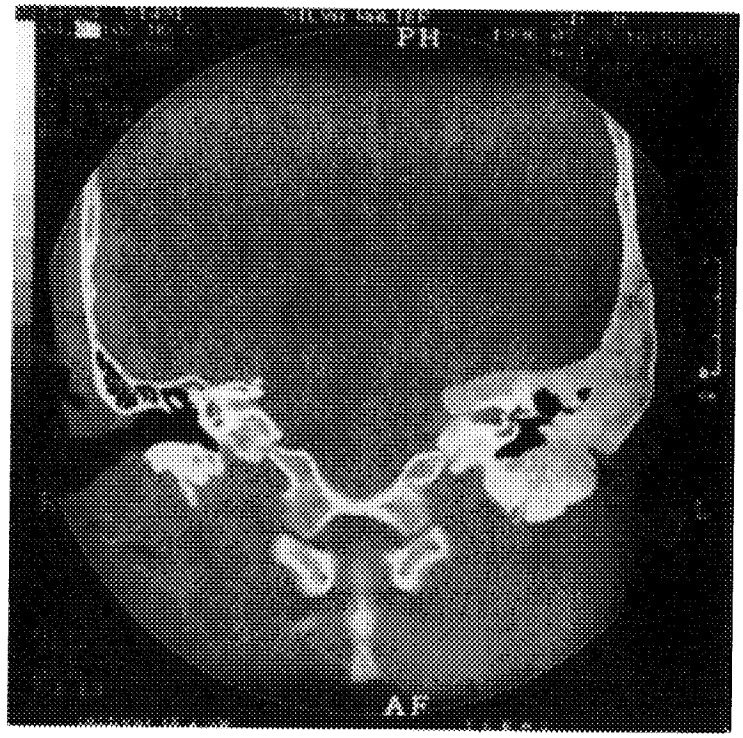


Fig. 4. Coronal CT scan: shows the stenosis of the external auditory canal of the left side.

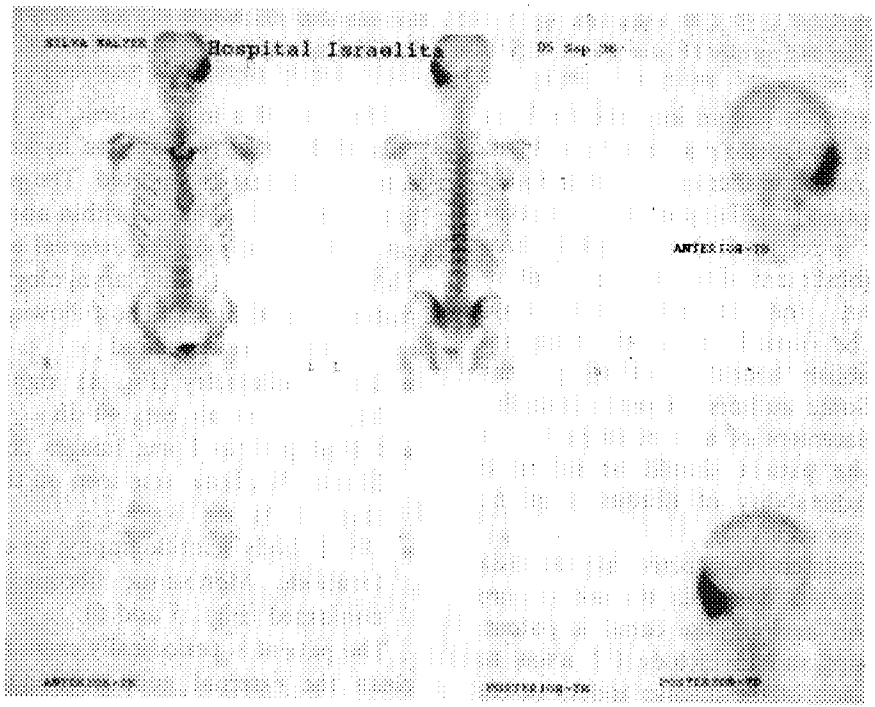


Fig. 5. Skelton scintillography shows an increase uptake of isotope Ga67 in the left side of the skull.

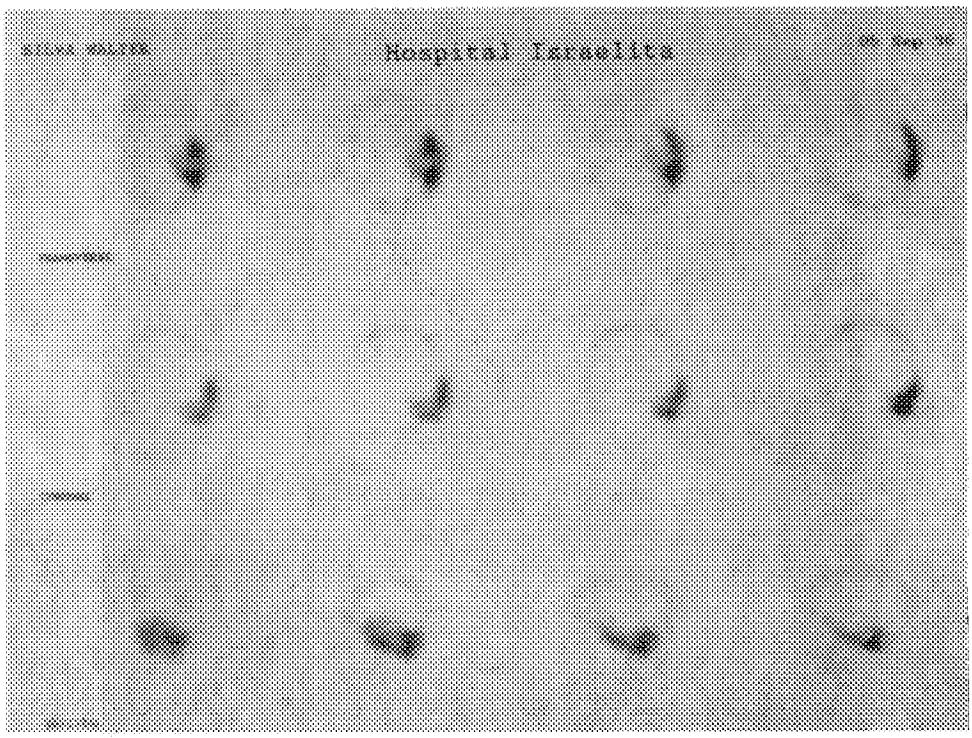

Fig. 6. Skull scintillography shows an increase uptake of isotope Ga67 in the left side of the skull.

## 7. Treatment

The use of corticoids has an important role in pain management [1]. Due to poor results and risk of malignancies, radiation therapy was abandoned as a treatment option [13]. Treatment is conservative and surgery is reserved to accomplish three objectives: reestablishment of function, prevention of complications, and cosmetic restoration. Surgery should be carried out in the temporal bone to re-establish hearing and to prevent cholesteatoma. Some authors advocate cannuloplasty [8] and placement of a stent to prevent a new stenosis. The patient should be informed about the unpredictability of fibrous dysplasia and its tendency to recur.

Patients with unilateral conductive hypoacousia resulting from fibrous dysplasia do not require surgical correction as long the canal is patent. Periodic follow-ups should include CT scans to evaluate progression of the disease and to detect the development of any secondary disorder.

## 8. Clinical case

The case of a male patient, 16 years old, who presented with a progressive hypoacousia in the course of 1 year is reported. The physical examination showed: right ear within normal limits, left ear with stenosis of the external auditory canal. The remaining otorhinolaryngological exam was within normal limits. The following complementary exams were requested:

- Tone audiometry (Fig. 1): right ear, normal; left ear, bone-air gap, 40 dB
- CT of petrous bone: images compatible with fibrous dysplasia were seen on the left mastoid (Figs. 2–4).
- Whole body scintillography: to eliminate other locations. Monostotic fibrous dysplasia was confirmed (Figs. 5 and 6).

The patient is periodically examined in order to clean the external auditory canal and remove cerumen. All progress is evaluated by CT.

## 9. Conclusions

Fibrous dysplasia is characterized by progressive replacement of normal bone by fibrous tissue.

The most common manifestation in the temporal bone is stenosis of the external auditory canal.

Treatment is conservative and surgery is reserved for preserving or restoring function and preventing complications.

Evolution is unpredictable.

Although this disease does not occur often, this diagnosis should be kept in mind for young patients with slightly progressive hypoacousia and the finding of external auditory canal stenosis during physical examination.

## Acknowledgements

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